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## Case Report

# Dyke-Davidoff-Masson syndrome: A case report with a literature review<sup>☆,☆☆</sup>

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### ABSTRACT

Dyke-Davidoff-Masson syndrome (DDMS) is an uncommon neurological disease defined as cerebral hemiatrophy with a contralateral motor deficit, facial asymmetry, and seizures. Classic imaging findings are cerebral hypoplasia, ventriculomegaly, paranasal sinus hyperpneumatization, and compensatory osseous enlargement. The diagnosis of DDMS is based on the correlation between clinical and neuroimaging features. The management of DDMS is based on anticonvulsant medication with physiotherapy. We describe an unusual case of DDMS presented with frequent and persistent seizures.

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## Introduction

Dyke-Davidoff-Masson syndrome (DDMS) is an atypical neurological disease described as cerebral hypoplasia or hemiatrophy, contralateral weakness, facial palsy, and seizures. Dyke, Davidoff, and Masson were the first to define this syndrome in 1933 [1]. It usually affects the pediatric population with unknown frequency.

The diagnosis of DDMS is based on the correlation between clinical and neuroimaging features. Classic imaging findings are cerebral hypoplasia, ventriculomegaly, paranasal sinus hyper-pneumatization, and compensatory osseous en-

largement [2]. Most clinicians misdiagnose and mismanage this neurological condition because of its rarity. We report a case of DDMS presented with recurrent seizures.

## Clinical case

A 10-year-old girl was admitted to our department 4 years after the onset of frequent right partial convulsions with occasional secondary generalization and walking difficulty. She was born as a full-term normal birth to nonconsanguineous parents with no pregnancy or postpartum problems. Her childhood development was normal.

<sup>☆</sup> Competing Interests: The authors have declared that no competing interests exist.

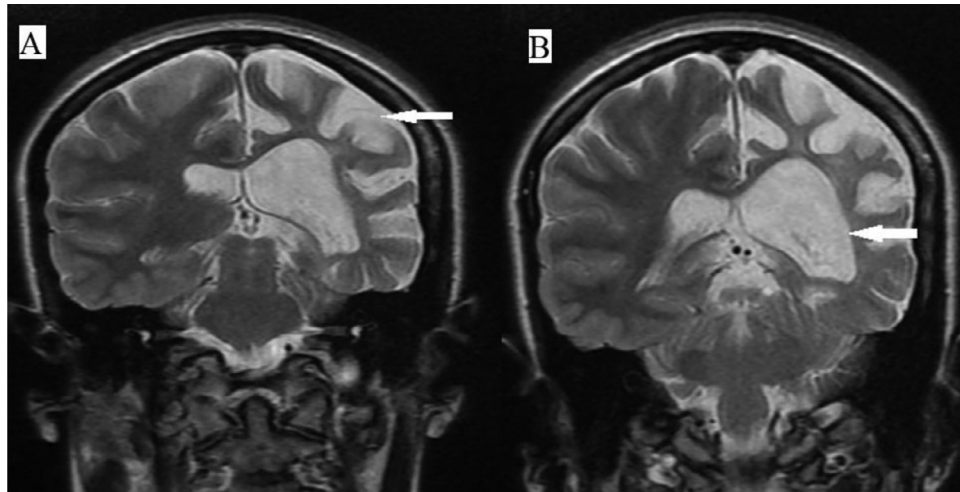
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**Fig. 1 – Cranial MRI on coronal T2 weighted sequence showing atrophy of the left cerebral hemisphere with prominence of the left Sylvian fissure (A), and dilatation of the left lateral ventricle (B).**

At the age of 6, she suffered from a fever for 3 days, accompanied by episodes of generalized seizures. The child was admitted and managed at a local health facility. The child continued to have seizures with a variable monthly frequency, generally caused by sleep deprivation, anxiety, and hyperthermia.

Gradually, she developed weakness on the right side along with speech difficulty. Later, the girl developed facial deviation, progressive right-sided weakness, slurred speech, and learning difficulty. She was not taking any medications.

On neurological examination, the patient had a right hemiparesis quoted as 3/5 on the Medical Research Council grading scale with a spastic gait. Deep tendon reflexes were brisk with plantar extensor response in the right side. She had a right hypoesthesia with right facial palsy and decreased cognitive functioning. There was no mucocutaneous lesion and the examination of other extra neurological systems was unremarkable.

Paraclinical tests, including full blood count, renal and liver function tests, blood glucose level, calcium, serum electrolyte, syphilis, and HIV test were normal. Investigations for young stroke with an immunological profile were unremarkable.

Brain MRI revealed left cerebral hemisphere atrophy, and ex vacuo dilatation of the left lateral ventricle with prominence of the left Sylvian fissure (Fig. 1). MRI showed also increased signal intensity in the left subcortical areas consistent with gliosis, hyper-pneumatization of the left frontal and sphenoidal sinuses, and compensatory calvarial hypertrophy (Fig. 2). EEG revealed epileptic abnormalities in the left hemisphere. We diagnosed this case as DDMS based on the history, clinical findings, neuroimaging and neurophysiological characteristics.

Sodium valproate was administered to the patient associated with physiotherapy. Although the dosage was increased to 25 mg/kg/d, the partial seizures persisted. Carbamazepine was added, and the frequency of seizures decreased.

## Discussion

Dyke, Davidoff, and Masson described for the first time in 1933, pneumoencephalographic abnormalities on standard skull radiography of 9 individuals with hemiparesis, facial asymmetry, seizures, and cognitive disorders [3].

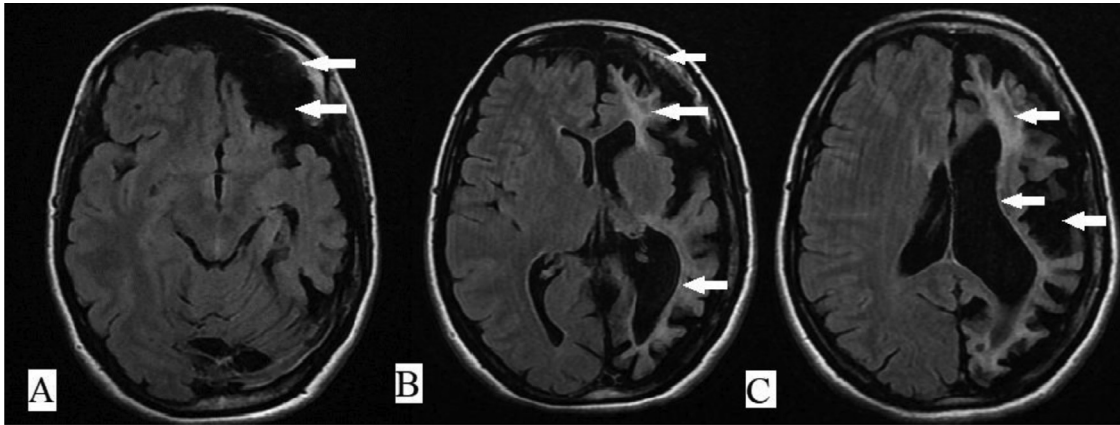
The participation of a particular hemisphere or gender predominance was not documented. However, male gender with involvement of the left hemisphere is more prevalent in the literature [4]. Partial or generalized seizures, contralateral hemiparesis, intellectual disabilities, learning problems, and poor speech are among the clinical manifestations of DDMS. The disease is categorized into 2 types based on its etiology.

The congenital form manifests itself in childhood, and its pathophysiology includes obstruction of fetal vascularization. The acquired form manifests in childhood. Perinatal hypoxia, infections, cerebrovascular abnormalities, and cranial trauma are among the etiological mechanisms [5]. The mechanism of cerebral hypoplasia and progressive neurological disorders is thought to be caused by a series of ischemia events due to various reasons, which limit the synthesis of brain-derived neurotrophic factors [6].

DDMS imaging findings include large cortical sulci, lateral ventricular dilatation, brain hemiatrophy, frontal sinus hyper-pneumatization, and compensatory skull thickening. As the child reaches maturity, these imaging results become more visible [7].

Sturge-Weber syndrome, brain tumors, Rasmussen encephalitis, and Fisherman syndrome are all differential diagnoses of cerebral hypoplasia. In most situations, a thorough clinical history, multisystem evaluation, and paraclinical investigation reveal an accurate diagnosis.

Since there is no defined protocol for managing DDMS, the treatment is mostly symptomatic, with anticonvulsive med-



**Fig. 2 – Cranial MRI on axial Flair sequence showing hyper-pneumatization of the left frontal and sphenoidal sinuses and compensatory calvarial hypertrophy (A), associated with left lateral ventricular dilatation and hyperintense signal in the left subcortical areas consistent with gliosis (B and C).**

ication used to control convulsions. Children with recurrent severe seizures and hemiplegia are prospective candidates for hemispherectomy, which has an 85 percent success rate in properly selected patients. If the motor impairment occurs after the age of 2 years with controllable seizures, the prognosis of the syndrome is favorable. Long-term patient management is based on physiotherapy, occupational, and linguistic therapy [8].

## Conclusion

DDMS is an uncommon condition that generates frequent and persistent seizures. Because it is a rare disease, physicians may easily misdiagnose it. Specific imaging findings associated with this syndrome can be revealed using MRI. To the best of our knowledge, this is the first case of DDMS reported from Morocco. In children with hemiparesis, facial asymmetry, or seizure disorder, it is critical to evaluate DDMS as a possible differential diagnosis so that the patients can be adequately managed.

## Patients consent

I qualify as the corresponding author to this manuscript warrant that I have informed the patient of this scientific manuscript and I confirm that I obtained his written and informed consent for the publication of this article.

## Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2022.04.047](https://doi.org/10.1016/j.radcr.2022.04.047).

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